Perioperative Management of Paediatric Patients With Genetic Diseases

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Paediatric patients with a genetic disease present a significant challenge for the surgeons, anaesthetists and nurses because of the significant comorbidities.

Many are preterm babies with congenital heart disease and compromised cardiovascular system. They may have pulmonary hypoplasia, reactive airway disease, chronic aspiration, recurrent pneumonia, chronic lung disease and need for long-term mechanical ventilation.

Neurologically; a significant proportion of patients have seizure disorder. Hypotonia is a common finding with delayed development and intellectual abnormalities. Some patients may have congenital hydrocephalus and they need insertion of VP-Shunt early in their life.

Metabolic and electrolytes abnormalities are common especially in patients with mitochondrial diseases, renal dysfunction and hepatic impairment.

Many patients have anaemia, poor nutritional status, global developmental delay and low weight for their age. All may increase the perioperative morbidities and mortalities.

As the patient may has limb contractures, spinal and skeletal deformities, care should be taken during positioning and pressure points should be protected.

Abnormal response to the medications and anaesthetic drugs is a common challenge. Careful selection of the drugs and precise calculation of the doses are paramount.

Regional anaesthetic techniques are usually recommended to reduce the doses of general anaesthetic medications and for easier management of postoperative pain. However the presence of a neurological disorder may preclude the use of regional techniques.
There is a greater chance for perioperative hypothermia, so all measures should be taken to minimise heat loss and to avoid hypothermia and its adverse effects.

Postoperatively; observation in high dependency unit may be needed for prolonged complicated procedures. Careful pain management is important as the child may suffer in silence. Control of postoperative nausea and vomiting will prevent aspiration and ensure a comfortable child.

The lecture will discuss the above challenges and draw a frame for proper perioperative care of this group of patients.